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ON

CONGENITAL MALFORMATIONS

OF THE

PALATE, FACE, AND NECK.

Three Demonstrations

GIVEN AT THE ROYAL COLLEGE OF SURGEONS, ENGLAND.

BY

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CONSERVATOR OF THE MUSEUM.

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Three Demonstrations

ON

CONGENITAL MALFORMATIONS OF PALATE, FACE, AND NECK.

I.

The material which has gradually accumulated in the various metropolitan medical museums during the last hundred years affords the basis for a very complete study of the various forms of cleft found in the palate and upper lip of children at birth. While the specimens which are shown at these demonstrations are derived from the museum of this college, it has also been possible for me, thanks to the unfailing courtesy of those in charge of the medical schools of London, to have free access to the valuable material on the shelves of their museums. A classified list of the total number of specimens at my disposal is given in Table I. Museum specimens are not representative of the run of cases met with in practice; the slighter and, from a surgical point of view, the more interesting lesions are sparsely represented, whereas the more severe forms, often associated with other grave lesions, predominate.*

Tripartite Palate.

In this, which must be regarded as the most severe form, the three elementary parts of the palate are widely separated. In all cases the lesion is remarkably alike, a typical example being shown in Fig. 1. In the middle element from before backwards are to be recognized (1) the middle part of the upper lip; (2) the premaxillary part of the palate, carrying the incisive papilla, which is joined to the upper lip by a fraenum; (3) the lower border of the

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* In 555 cases at the clinic of von Bruns at Tübingen, Dr. Gustav Haug (*Beiträge fur klin. Chirurgie*, 1904, Bd. 44, p. 254) found that the tripartite palate occurred in 15 per cent. (against 38 per cent. in the London series), bipartite palate in 23 per cent. (against 22 in the London series), unilateral harelip with no cleft of the palate 40 per cent. (against 9 per cent. in the London series).

septum of the nose, which is wider, shallower, and longer than normal; (4) the varying number of dental sacs on the premaxilla: in 16 cases only the two middle incisors are carried; in 3 cases one lateral incisor as well as the two middle (as in the specimen shown in Fig. 1), and in 4 cases all four incisors. In no case did the premaxilla in these cases show a suture between the ectognathic and mesognathic parts. The nerves and vessels of this element are the naso-palatine. On each lateral or maxillary element three distinct areas can be recognized (Fig. 1): (1) the alveolar area on which may be seen an elevation over the sacs of the two milk molars, another over the canine and frequently a third, often on the margin of the cleft, over the lateral incisor (Fig. 1); (2) a plicate or rugose area marked by folds; (3) a smooth narrow marginal area (see Fig. 1). When the mucous membrane is removed, the maxillary or lateral element is seen to be supported by the superior maxilla and palate bones. The nerves and vessels of the maxillary elements are the descending palatine.

TABLE I.—*Classified List of Specimens showing Congenital Clefts of the Palate and Upper Lip in Metropolitan Medical Museums.*

	A.	B.	C.	Total.
1. Palate cleft into three parts	14	12	2	28
2. Palate cleft into two parts	3	12	2	17
3. Median cleft extending from the premaxilla to the uvula	4	3	7	14
4. Cleft of soft palate	3	0	0	3
5. Cleft of uvula	4	1	0	5
6. Cleft on both sides of premaxilla	1	0	0	1
7. Cleft limited to one side of premaxilla ...	1	0	1	2
8. Median cleft due to absence of premaxilla	1	2	4	7
9. Bilateral harelip	1	0	3	4
10. Unilateral harelip	0	6	0	6
11. Median harelip	—	—	—	0
	32	36	19	87

A. Specimens in the Museum of the Royal College of Surgeons of England.
B. Specimens in the museums of metropolitan medical schools.
C. Specimens of cleft palate in mammals other than man, chiefly in the Museum of the Royal College of Surgeons of England.

The Nature of the Lesion.

Seeing that fusion of the three elements to form the palate begins in the human embryo at the fifth week, a period at which the branchial arches are best marked, it is only natural that one should turn to fishes to find as a normal condition one comparable to that seen in the tripartite

palate of the malformed child. Embryology, too, leaves us in no doubt as to the class of fishes in which the condition should occur; the more we know of the early development of the mammalian embryo, the more we become convinced that it has much in common with the Selachii or shark tribe. The tripartite palate is an arrest of development at a Selachian stage. In Fig. 2 is shown the mouth of the dogfish; what at first sight appears to be its upper lip is divided by two furrows—the olfactory or naso-buccal—into three parts, a mesial and two lateral. The part between the furrows represents the middle nasal process of human embryology—the process which gives rise to the nasal septum and premaxilla. The furrows

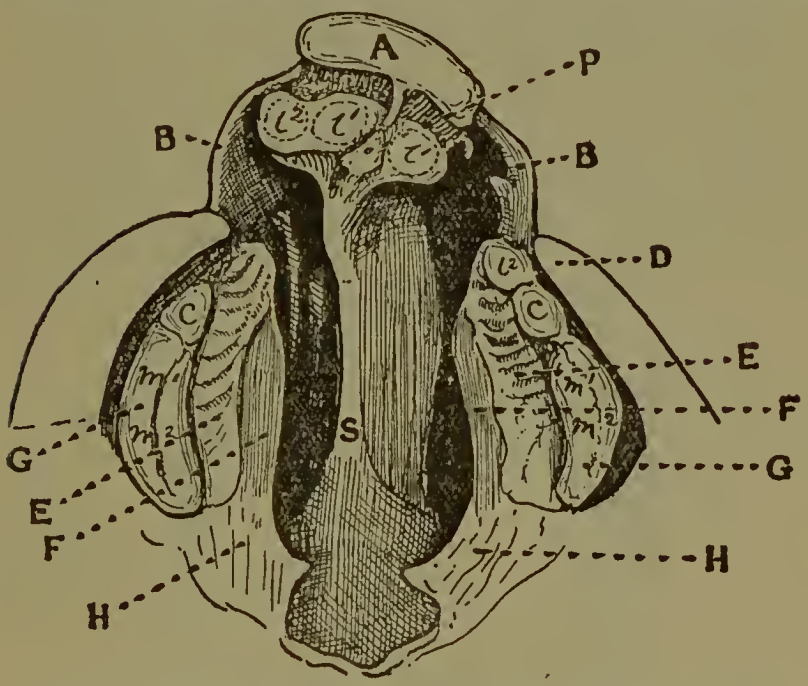

Fig. 1.—Tripartite palate. A, Median part of upper lip; B, C, alae of nose (two tags on left); C, canine tooth buds; D, lip; E, rugose part of palate; F, marginal part; G, alveolar part; H, soft palate with uvular process; I^1, I^2, incisors; P, incisive papilla; S, septum.

which represent extensions of the nasal openings towards, or actually into, the buccal cavity of the shark, are divided by a fold from the middle process (see Fig. 2) into two functional parts—an upper, the anterior nares, by which water may enter the nasal cavity, and a lower or posterior nares, by which the water may be drawn into the pharynx as that cavity expands during respiration. At first sight the comparison seems imperfect, for the nasal furrows of the shark appear to lie rather on what may be named its face and entirely in front of the teeth. But these teeth behind the furrows in the shark's mouth are carried not by the maxillae but by the palate bones—bones repre-

sented in the human skull by the vertical processes of the palatines—situated, as is the shark's, at the posterior nares. The premaxilla and the premaxillary part of the palate are absent in the shark, but there can be no doubt that the area of tissue lying between the nasal furrows represents the part which bears the premaxilla in higher vertebrates, and that in the course of evolution the roof of the mouth has been extended on to this area.

In those remarkable air-breathing fishes, the Dipnoi, the margins of the nasal furrows have actually fused. The bridge of tissue thus formed across the nasal furrows is the earliest form of union between the mesial and lateral elements of the palate. The condition of cleft palate, normal in the shark, has disappeared in the Dipnoi, and all vertebrates higher in the scale than fishes.

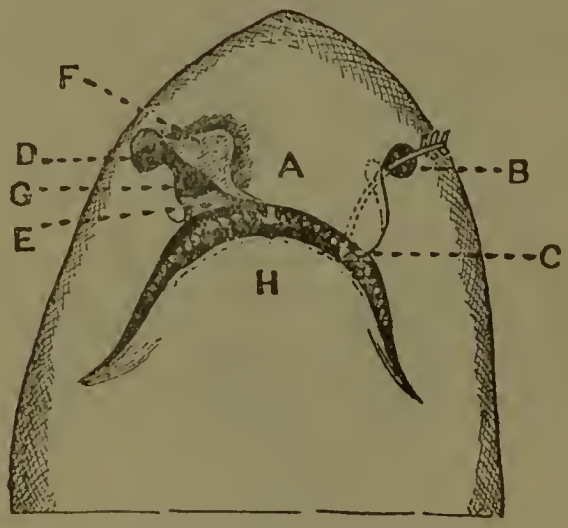

Fig. 2.—Head and mouth of a young dogfish (*Scyllium canicula*). A, Internasal field; B, anterior nares; C, posterior nares; D, anterior nares and olfactory pit; E, posterior nares; F, opercular fold (turned up), which separates the anterior nares from the posterior; G, maxillary process; H, lower jaw.

The stage of development seen in the Dipnoi may be studied more conveniently in amphibians. In Figs. 3 and 4 are shown side by side the palate of one of the larger frogs and of a lion cub—one of the many which have been born in the gardens of the Zoological Society of London with cleft palate. In the frog's palate a premaxilla has appeared, and it has extended across the nasal furrow and fused with the maxilla, so that a horseshoe-shaped alveolar arcade is now formed. The amphibian form marks a distinct stage in the evolution of the palate, and one at which development may be arrested, as shown by the specimen represented in Fig. 4. In the human embryo the Selachian stage is superseded by the amphibian during the fifth and sixth weeks of development; the mesial or premaxillary processes unite then with the lateral or maxillary element of the palate. During the

seventh and eighth weeks the third or mammalian stage is entered on; from the lateral or maxillary elements two palatal processes grow out and fuse in the middle line, fusion commencing behind the premaxillary element and ending at the uvula (Fig. 4A). Within the palatal processes the horizontal plates of the maxillary and palate bones are developed. Arrest may occur at any point of the third stage. The development of the horizontal palate rendered the evolution of mammals possible; without such a palate suction and mastication are impossible, unless the breath be held while these acts are in progress.

The Degree of Separation between the Various Parts of the Palate.

The late Professor His was of opinion that clefts in the palate resulted from irregular and inco-ordinated growth of the three elements which go to form it. That the growth of the three parts is irregular in the later months of fetal life there can be no doubt (see Figs. 1 and 5). The septal part of the mesial nasal process is abnormally long, so that the premaxilla projects some 6 or 8 mm. in

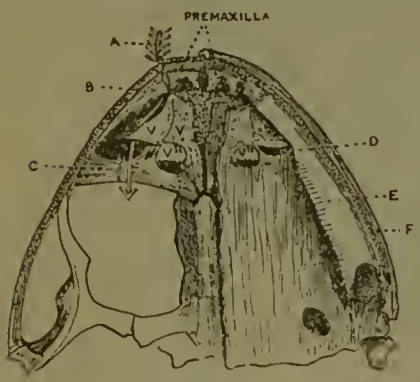

Fig. 3.

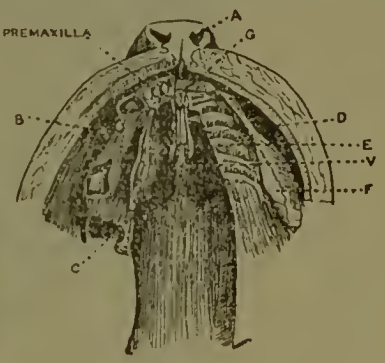

Fig. 4.

Fig. 3.—Palate of frog. Fig. 4.—Palate of lion cub with cleft palate. In both figures the mucous membrane is removed and the bones exposed on the right side. A, Anterior nares; B, palatal process of maxilla; C, palate bone; D, posterior nares; E, rugose part of maxillary palate; F, alveolar palate; G, incisive papilla with organ of Jacobson opening on each side of it; V, vomer; V″, vomerine teeth.

front of the maxillary parts of the palate. The increase in length of the septum is due not to the vomerine part of the septum, but to that part of it which is formed by the premaxillary processes (see Fig. 7). If the premaxilla becomes adherent to the maxilla on one side the extra growth of the septum still takes place, with the result that the premaxilla is bent towards the side on which union takes place (Fig. 5). Irregularity of growth is not the cause but the result of the cleft condition. From the evidence to be produced it is probable that the processes which form the palate were in contact during the fifth and sixth weeks of development, but for some reason—the exact cause we do not know—union did

not take place then. The union of embryological processes is in every way comparable to the healing of wounds; the epithelial coverings on each side of the nasal groove come in contact and form a bridge across which the uniting mesoblast may grow. If for some reason union is delayed—and from Professor Mall's observations there can be no doubt that uterine inflammation, sometimes from syphilis, is the most common cause—growth in the several elements of the palate causes them to separate, and once a breach in their continuity has been effected, union cannot afterwards take place. The younger the fetus the smaller the cleft, not only absolutely but relatively. The width of the fissure between the palatal processes of the superior maxilla varies from 7 to 20 mm. in the tripartite palate; 14 mm. is the average in eight full-time children; in a fetus at the fourth month this cleft measured only 1 mm. Brophy[1] has suggested that the subsequent separation is

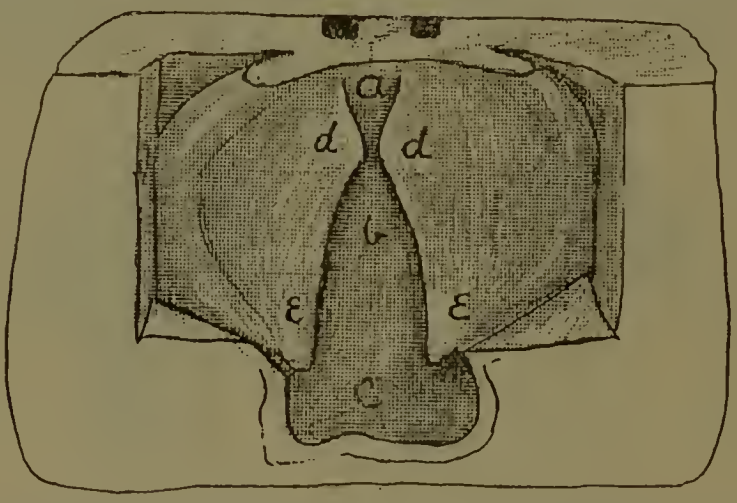

Fig. 4A.—Model of the developing palate of a human fetus about 6 weeks old (28 mm. long) (Anna Pölzl). *a*, Premaxillary part of palate; *b*, septum of nose; *c*, naso-pharynx; *d d*, maxillary plates about to meet; *e e*, soft palate and uvula.

caused by pressure of the lower jaw and tongue against the palate, and the material at my disposal is in harmony with his suggestion. He also makes the statement that there is no atrophy of the parts that go to form the palate, but this is certainly not the case in the specimens I have examined. In three palates of adults the cleft was in the average 20 mm. wide, the breadth of the palate 64 mm.; the bony parts of these palates were thus 10 or 15 mm. less than normal. In newly-born children the deficiency affects the inner or marginal area of the palate (see Fig. 1), but it does not amount to more than 3 mm. on each side. Whether the cleft continues to increase after birth I am not in a position to say, but, judging from the width of the cleft in the adult palate, I conclude that the parts do not continue to separate after birth at the rate they did before birth.

Imperfect Degrees of Union.

In Fig. 5 there are shown very imporfect strands of union between the pre-maxillary and maxillary processes. A study of these throws much light on the condition of cleft palate. The slightest degree of union is shown in Fig. 5A, where a strand of fibrous tissue covered by epithelium unites the ala of the nose to the premaxillary process; this represents the rudiment, or rather vestige, of the union of the mesial and lateral nasal process to form the boundary of the anterior nares. In B, C, D (Fig. 5) increasing degrees of union are shown. In the fullest degree (Fig. 5, D) the uniting band forms a bridge on which four elements end in attempting to reach the pre-

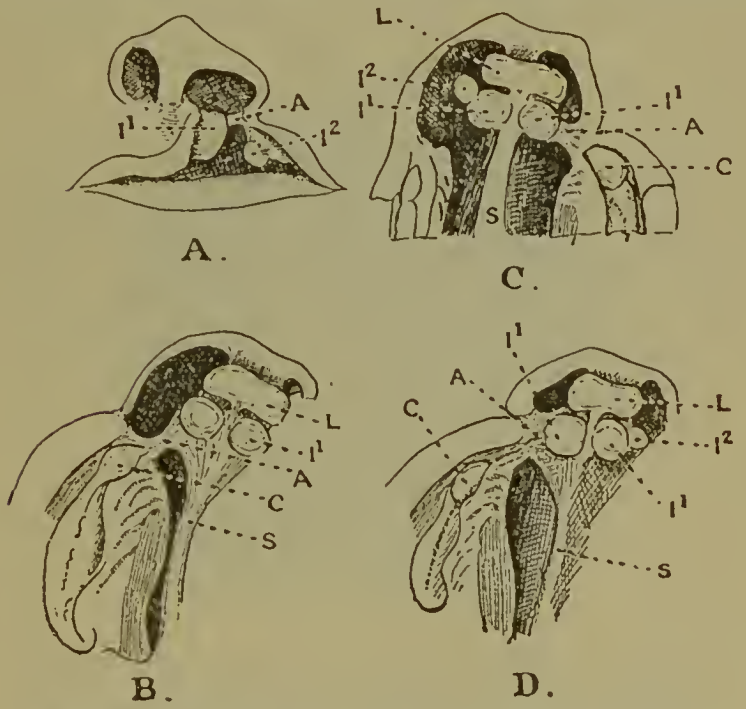

Fig. 5.—Four specimens of cleft palate showing various degrees in the development of the bond between the premaxillary, maxillary, and lateral nasal elements. A, The bond or bridge of tissue crossing the cleft; I^1, central incisor sac; I^2, lateral incisor sac; C, canine sac; L, median part of upper lip; S, septum of nose.

maxillary process—(1) the ala of the nose, (2) the lateral part of the upper lip, (3) the dental groove, (4) the palatal process of the upper maxilla. These strands of union have apparently become stretched by the unequal or irregular growth of the parts which go to form the palate and upper lip; occasionally blunt conical processes occur on each side of the cleft, evidently the remnants of a strand which has broken under the strain.

The Relationship of the Incisor Teeth to the Clefts.

It is well known that the cleft usually lies between the middle and lateral incisors. In 41 cases I was able to

verify the position of the fissure; in 23 it passed between the lateral and middle incisors; in 9 it passed between the lateral incisor and canino. In 7 cases the lateral incisor had not developed on the side of the fissure; in 2 cases a third incisor was developed. To explain the varying relationship of the fissure to the incisor teeth Albrecht supposed that the premaxilla was developed in two parts, that carrying the mesial incisor being developed in the mesial nasal process, and that carrying the lateral, in the lateral nasal process. Koelliker, and afterwards His, showed conclusively that Albrecht was wrong—that the lateral nasal process did not enter into the formation of

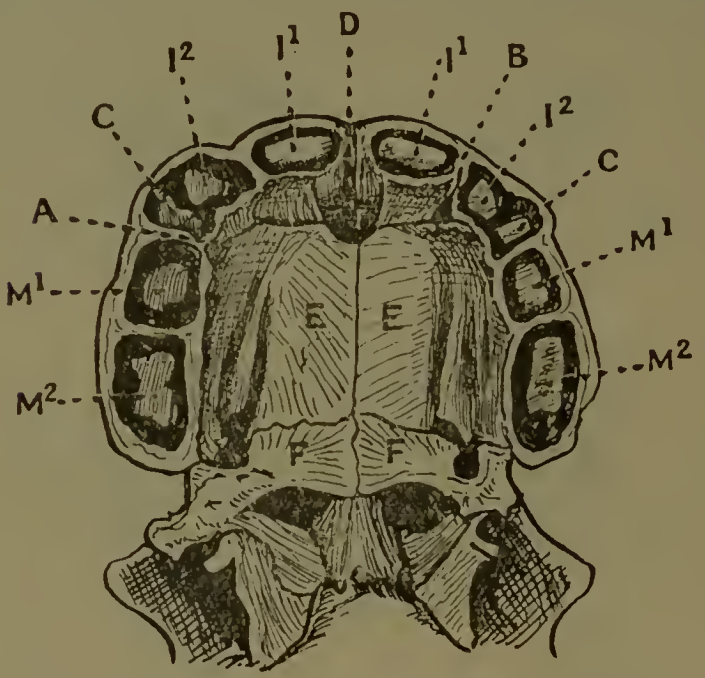

Fig. 6.—Palate of newly-born child (natural size) to show the elements entering into its formation. A, Suture between premaxilla and maxilla ending behind canine tooth; B, the same ending behind central incisor; C, canine; D, vomer appearing in naso-palatine fossa; E, marginal part of maxillary palate; F, horizontal process of palate bone.

either the palate or lip, but because Albrecht offered an explanation of appearances with which surgeons are familiar while the great anatomists offered none, his theory has been widely accepted in England. The true explanation of the varying relationship of the teeth to the fissure is extremely simple. The germ of the lateral incisor, although carried by the mesial nasal process, is laid down in the cleft between the maxillary and pro-maxillary (mesial nasal) processes. In cases of cleft palate the processes move apart under the strain of growth during the middle and later months of fetal life. Three fates may then overtake the bud of the lateral incisor: it may be destroyed, it may remain attached to the pro-

maxillary process, but more frequently it moves outwards attached to the maxillary process. I have seen it stranded on the bridge of tissue between the processes or loosely attached at one side of the fissure or the other (see Fig. 5, c). Mr. Clement Lucas has pointed out that families in which harelip and cleft palate are hereditary have usually the lateral incisor of very small size; Professor Warnekros regards an irregularity in the development of the lateral incisor as the actual cause of cleft palate, and points out not only the frequent absence but the occasional duplication of this tooth in families in which cleft palate is apt to occur. Yet the bud of the lateral incisor belongs to and is developed in connexion with the mesial nasal process. In the condition of cyclops, where this process fails to take part in the formation of the palate, all the incisor teeth are absent.

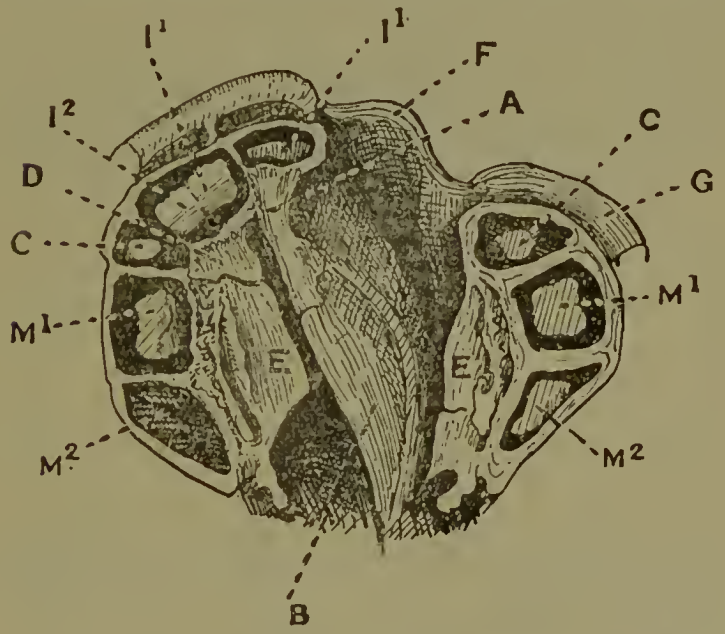

Fig. 7.—Bipartite palate. A, Septal process of premaxilla; B, vomer; C, canine; D, premaxilla; E, marginal part of palate; F, ala of nose; G, maxillary part of lip; I^1, I^2, incisor teeth; M^1, M^2, milk molars.

Abnormalities in the Ossification of the Premaxilla.

Albrecht called attention to the fact that there is often a double centre of ossification in each premaxilla. (See Fig 6.) It will be observed that the amphibian premaxillae are subdivided (Fig. 3). The position of the suture between the premaxilla and maxilla and its relationship to the teeth are extremely variable. As a rule it ends between the lateral incisor and canine, but it may vary in position, as Schumacher has pointed out, to the degree illustrated in Fig. 7, ending in extreme cases either between the lateral and middle incisor or between the canine and first milk molar. The lateral part of the premaxilla is formed within the maxillary process; the process of ossification is

not influenced by the position of embryonic fissure; the premaxilla is laid down in adjacent parts of all three processes that go to form the face.

The Bipartite Palate.

Having discussed in some detail the tripartite palate it will be sufficient to deal with the less severe, if commoner, forms more briefly. In the bipartite palate the premaxillary element has undergone normal union on one side, usually the right, with the maxillary process (Fig. 7). Of the 15 cases in man, the cleft passed to the left side of the premaxilla in 11; Haug found it on the left side in 149 cases out of 216. Why the left side is the more frequently cleft I cannot say. The nasal septum in such

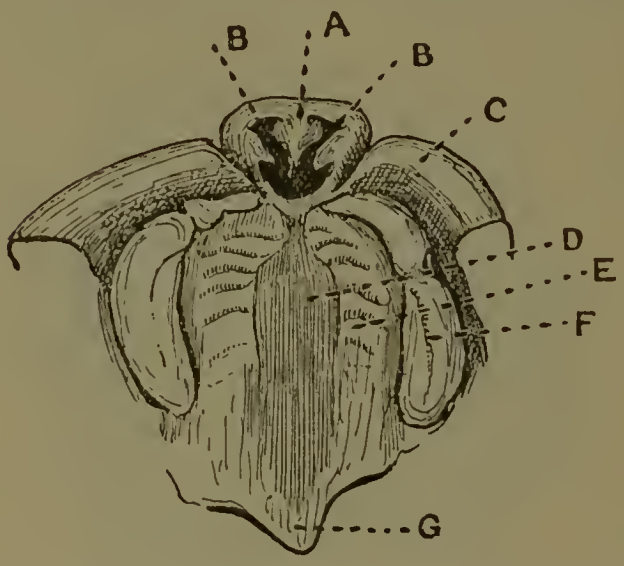

Fig. 8.—Absence of premaxillary part of palate (preparation No. 203). A, Vestige of septum and premaxillary process; B, B, anterior nares; C, lateral part of upper lip; D, marginal part of maxillary palate; E, F, rugose and alveolar parts; G, soft palate and uvula.

cases is always markedly deformed, presenting its convexity to the side on which the cleft is situated. In 4 of these cases union had occurred not only between the premaxilla and maxilla, but also between the nasal septum and edge of the palatal processes of the maxilla and palate bone (see Fig. 7). In the usual form the lower border of the nasal septum is free, there being no union between the maxillary and mesial nasal elements.

Intermaxillary Clefts.

As we have seen, the union of the premaxillary element with the alveolar parts of the maxilla marks a distinct stage in the evolution of the palate—a stage well seen in amphibians. The next stage—the mammalian—lies in the development and union of the maxillary palatal processes already to be seen in the amphibian palate

(Fig. 3). One would expect arrests to occur in the passage from the amphibian to the mammalian stage (see Fig. 3), although such specimens are comparatively rare in museums (of the 14 cases on my list, 7 are from the human subject, the others are from the dog, lion, sheep, and calf); they are the more common form of cleft palate met with in practice. Mr. J. Berry[2] records a list of 67 cases, of which 36 were due to non-fusion of the maxillary processes in the roof of the mouth. In these cases there is a real arrest, not only of the fusion of the processes, but of their actual growth. Clefts of the soft palate alone or of the uvula alone, as may be seen from Table I, are comparatively rare; I have seen 3 of the former and 5 of the latter. They indicate an arrest towards the end of the development of the palate when the fetus is about 50 mm. long and entering the third month of intrauterine life. Even when the palate is completely cleft the two halves of the uvula are clearly indicated as tags projected from the border of the palatal folds. The dissections in the College Museum show the tensor and levator palati well developed; the upper part of the superior constrictor and palato-pharyngeus are stronger than in normal palates.

Premaxillary Clefts.

The cleft or clefts may be limited to the premaxillary part of the palate. In the three cases included in Table I the fissure was unilateral in two cases and bilateral in the third. In these cases arrest had occurred during the period of formation of the primary palate, whereas the process of formation of the secondary palate had been successfully carried out. Besides lateral premaxillary clefts, there are two others, mesial in position, which are rare but worthy of notice. In one of these forms the embryological elements (the globular processes) which go to form the middle third of the upper lip and premaxillary palate have been arrested at an early stage, and a mesial cleft is thus left in the lip and palate. Of the 7 cases on my list 3 are from human subjects (see Fig. 8) and 4 from pigs. In such cases a lateral incisor may be present. The other form is very rare—namely, where a mesial cleft occurs in the upper lip similar to that which occurs in the hare. Cases of this cleft are described by Mr. Clutton and Mr. Edmund Owen in man. The two globular processes have not completely united together. This anomaly is much commoner in the domestic mammals than in man, as shown by the specimens in the College Museum.

The Causation of Harelip.

Although comparatively few family histories are at present available, there is a general agreement among those who have inquired into the matter that harelip and cleft palate are very apt to occur in certain families. Mr. Edmund Owen, Mr. Clement Lucas, and Professor Warnekros have recorded family trees showing the degree

to which the condition may be inherited.* More suggestive of the true cause of the condition are (1) the malformation with which cleft palate is so often associated (spina bifida, anencephaly, morbus cordis, atresia ani, and club-foot); (2) the observations of experimental embryologists. Such malformations may be produced by disturbing the development of the embryo by various means, but one may conclude from Professor Mall's observations on malformed human embryos that morbid uterine or placental conditions are the commonest cause of malformation of the lip and palate in man. In a group of sixteen malformed human fœtuses, the prevailing lesion being anencephaly, the following defects of the palate were found: Divided uvula 4, divided soft palate 1, division of hard and soft palates 1, anterior bilateral fissure 1, unilateral premaxillary fissure 1, bipartite palate 1, failure of ossification of hard palate 1. Eleven out of the sixteen showed malformation of the palate. These malformations are rarely met with amongst natives of Africa and of the South Sea Islands.†[3]

REFERENCES.

[1] Brophy, T. W., *Jour. Amer. Med. Assoc.*, 1907, vol. xlix, p. 662. [2] BRITISH MEDICAL JOURNAL, October 7th, 1895. [3] Murray, R. W., *Lancet*, 1904, vol. i, p. 1423.

* For details as regards heredity see Le Dentu. *Comptes rendus de l'Académie des Sciences*, 1908, t. 146, p. 1138.

† A skull of a South Sea Islander in the college collection shows a cleft of the palate.

II.

Congenital Fistulae of the Lower Lip.

A bilateral cleft in the upper lip is frequently associated with a curious malformation of the lower lip, in which two fistulae or recesses open on the lower labial surface opposite the clefts in the upper lip. The orifice of each recess may be raised into a nipple-like process, which, when the mouth is shut, fits into the corresponding upper cleft. This was the condition in a specimen submitted to me by Mr. Woolcombe,[1] which was excised from the lip of a boy, aged 3 months. The orifice of the recess was situated at the apex of the papilla; its fundus, about the size of a rice-grain, lay within the substance of the lip, and was surrounded by mucous glands which opened into it. A layer of striated muscle surrounded the pocket, which in nature was clearly a localized invagination of the mucous membrane and glands of the lip. In three cases reported recently by Mr. R. C. Dunn,[2] there was one in which the recesses opened on nipple-like processes; another in which the orifices were wide and flush with the surface of the lip; while in the third the two recesses were confluent—thus forming a transverse depression on the surface of the lower lip. The condition has been described by Mr. Bland-Sutton, Mr. Arbuthnot Lane, Mr. Clutton, and Dr. Ballantyne. There is no example of this malformation in London museums. An appeal to comparative anatomy does not afford a satisfactory explanation. Seeing that the malformation is so closely associated with bilateral harelip and cleft palate, a condition which occurs normally in certain fishes, it is clearly in this vertebrate class that an explanation of the condition is to be sought. On each side of the middle line of the lower lip of sharks, and exactly in the position where these recesses are found, open a group of mucous canals which are connected with nerve endings found in fishes. It is possible that these labial recesses have some relationship with these two mucous labial organs found in selachians, but the matter requires further investigation.

Median Cleft of the Lower Lip and Mandible.

Among the 250 specimens of malformation examined, only 4 showed this condition—a full-time child in the museum of St. George's Hospital and three specimens in the museum of this college, one from an ass, another from

a cockatoo, and a third from a sparrow. Seeing that the lower lip and mandible arise by the fusion of right and left halves, their immunity from median fissure, as compared with the upper lip and palate, is surprising. In four other specimens there was an apparent cleft of the lower jaw, but when the condition was more minutely examined it was seen that the lesion was really the result of a bifurcation or doubling of the buccal cavity, an attempt at the formation of twins.

TABLE II.

	A.	B.	C.*	Total.
Recesses and papillae on the lower lip ...	—	—	—	—
Median clefts on the lower lip and mandible	—	1	3	4
Naso-maxillary cleft	—	4	—	4
Lateral nasal clefts...	3	1	—	4
Mesial nasal clefts	—	—	5	5
Lateral nasal proboscis	—	—	—	—
Recess on nasal septum	—	—	—	—
Congenital perforation of nasal septum ...	1	—	—	1
Occlusion of anterior nares	2	—	—	2
Occlusion of posterior nares	—	—	—	—

* For explanation see Table III.

Naso-maxillary Clefts.

In four fetuses, all of them nearly at full time, the maxillary process is widely separated from the lateral nasal process, so that the nasal duct is represented by a wide groove, lined by mucous membrane, and shelving from the inner angle of the eye into the nasal cavity (Fig. 9). These specimens are in the museums attached to the medical schools of St. Bartholomew's, St. Thomas's, and the "London" Hospitals. In two the cleft is on the right side and in two on the left. The nasal duct and lacrymal canaliculi are formed at the same period of development as the primitive palate—namely, during the sixth and seventh weeks, when the maxillary and lateral nasal processes fuse across the intervening groove and enclose a rod of solid epithelium between them. The rod becomes canaliculized early, but, according to Stanguleanu[3], its nasal end does not open until the end of the eighth month. In all four specimens there was double harelip and cleft palate. In the specimen figured the cleft had evidently been continued into the temporal region on the right side, a small half-submerged polypus marking its termination on the right temple. The nasal duct appeared during the evolution of the amphibia, but in no living animal is any stage known between the complete formation of the duct as seen in amphibia, and the open skin area be-

tween the nose and eye as seen in sharks. There are no specimens in the metropolitan museums which illustrate the occurrence of dermoid cysts along the naso-maxillary fissure. Ballantyne met only three specimens of fissure in the cheek in 325 cases of congenital malformation.

Lateral Nasal Clefts.

Through the courtesy of Mr. Raymond Johnson I was lately afforded an opportunity of examining a child which showed a very uncommon and very interesting malformation of the nose (Fig. 10). There was a complete cleft of the palate, the cleft ending anteriorly in the left nostril. The wing of the left nostril (lateral nasal process) is pulled outwards and is widely separated from the septal

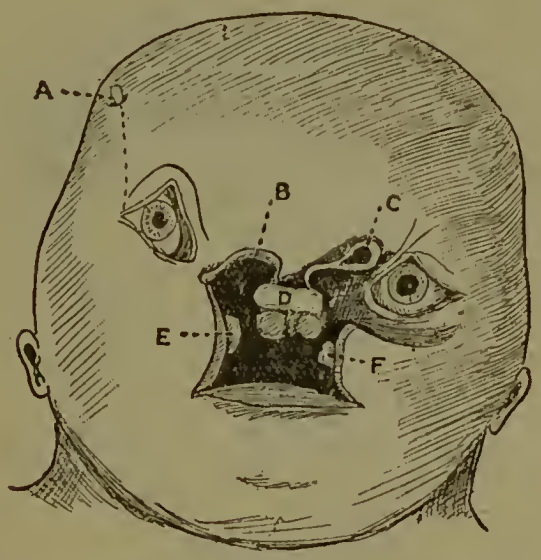

Fig. 9.—Full-time child, showing tripartite palate and naso-maxillary cleft. A, Polypus in temporal region, probably connected with a naso-maxillary cleft; B, C, right and left lateral nasal processes; D, mesial nasal process; E, F, right and left alveolar maxillary processes.

part of the nose (the mesial nasal process). The cleft thus lies between the mesial and lateral nasal processes on the left side. On the forehead there is an interfrontal depression covered by hair which descends towards the nasal cleft. There is no specimen exactly like this in the metropolitan museums, but there are four—three of them in the museum of this college—which help us to understand the condition. It is one due not to an arrest of the normal progress of development, but to certain bands, probably amniotic in origin, which are apt to occur in certain pathological conditions of development between the anterior nares or lips and the scalp. Professor von Winkel,[4] who regards most facial malformations as a result of amniotic adhesions, has figured a case in which such a band passed from each nostril over

the forehead to end in spongy tissue on the scalp. In two specimens in the college collection the scalp and calvarium are represented by a spongy vascular mass of tissue; a deep furrow passes from this mass along the nose to a premaxillary cleft on the left side of the palate (see Fig. 11). The left ala of the nose bounds one side of the fissure in which the left nasal cavity is exposed. The right anterior naris is perfectly formed, for the furrow, as in Mr. Raymond Johnson's case, lies between the mesial and lateral nasal processes of the left side. In another specimen in the college collection epidermal filaments—adhesive cords, such as are regarded as amniotic bands—project from and close the nostrils, but the face is otherwise well formed. In a specimen of

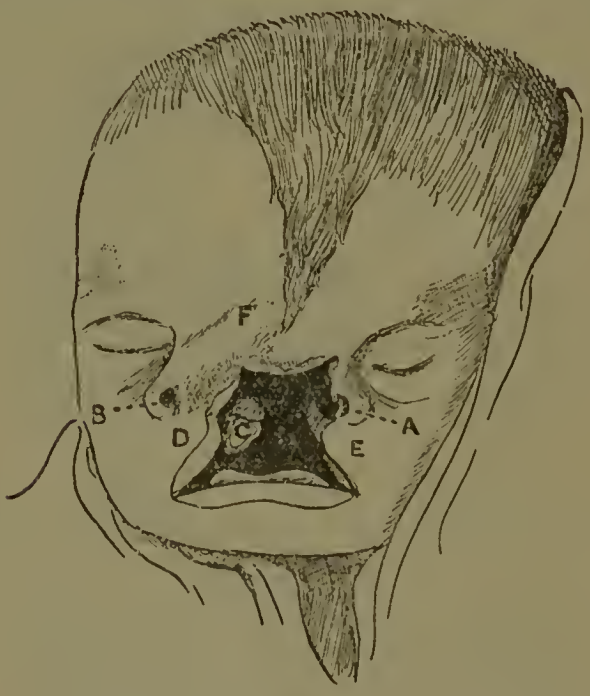

Fig. 10.—Child showing nasal cleft with depression on forehead (Mr. Raymond Johnson's case). A, Ala of left nostril; B, ala of right; C, sacs of incisor teeth; D, mesial, nasal, and right maxillary elements; E, left maxillary element; F, depression on forehead marked by prolongation of scalp.

anencephalic fetus in the museum of University College Hospital Medical School the nostrils are small and closed; from the root of the nose bands of tissue, apparently amniotic in origin, are attached. Taking all these cases together, one may reasonably infer that in Mr. Raymond Johnson's case (Fig. 10) an abnormal band of epidermal or amniotic tissue had been formed at an early stage of development between the coverings of the cerebrum and buccal cavity, the depression in the forehead indicating the position of the band. Why bands should be formed so exactly in the

same position in case after case requires further elucidation. We shall see presently that fissures of a similar nature occur between the mouth and the ear.

Mesial Nasal Fissures.

There is not a single museum specimen in London to illustrate the occurrence of a fissure in the mid line of the nose in the human subject, nor is there a specimen of the dermoids or of the fistulae which occasionally occur in the line of this fissure. Dr. Alex. MacLennan [5] has recently reported a case in which a tumour, dermoid in nature, occupied an area corresponding to the flat, wide interolfactory field of the human embryo and of the shark, and had prevented the proper apposition and fusion of

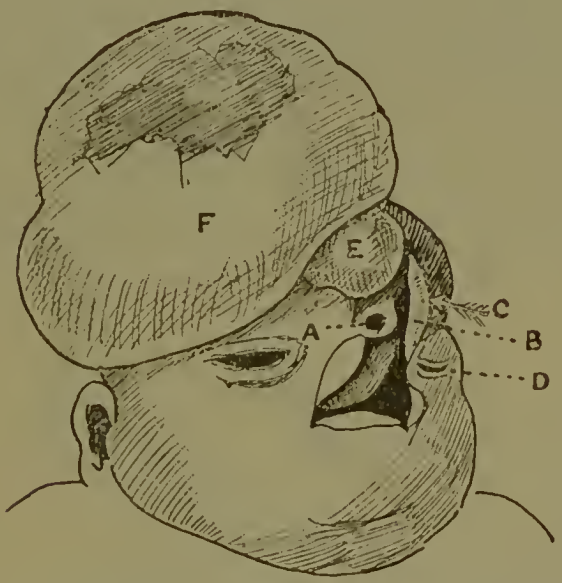

Fig. 11.—Full time child showing a cleft in nose, probably caused by an amniotic band. A, Right nostril; B, ala of left nose, bounding left side of a fissure which leads up to the masses of tissue E, F; C, the arrow points to the left nostril; D, left eye. A condition of exomphalos was also present.

the symmetrical right and left halves of the nose. An incomplete fusion in the mid line of the two halves of the external nose is comparatively common amongst animals; five specimens in the college museum illustrate the condition—three from the pig, one from the cat, and one from the dog. Mr. Bland-Sutton has drawn attention to a breed of Parisian pug dogs in which a mesial nasal fissure forms a constant feature.

Lateral Nasal Proboscis.

In certain rare cases one half of the nose is arrested at an early stage of development and assumes the form of a small proboscis, such as is shown in Fig. 12. Some eight years ago Mr. Frederic Eve gave me the opportunity of

examining a boy at the London Hospital in which this abnormality was present, the proboscis being situated over the inner canthus of the right eye in place of over the left canthus, as in Fig. 10. In many specimens of cyclops the nose is represented by such a proboscis (see Fig. 15); a lateral nasal proboscis represents a half—right or left as the case may be—of the proboscis seen in a cyclops. Mr. Eve removed the proboscis, and it was found to consist of a central pocket of mucous membrane, encapsuled in a thin plate of cartilage and covered by skin. Mucous fluid escaped from it, as in Kirchmayer's case, when the boy cried. The epithelial lining of the mucous pocket is stratified squamous at the trumpet-shaped entrance, and columnar in the rest of its extent. The primary lesion in these cases appears to be a unilateral absence or failure of the epithelial and nervous substratum of the organ of smell; the stimulus which gives rise to the full formation

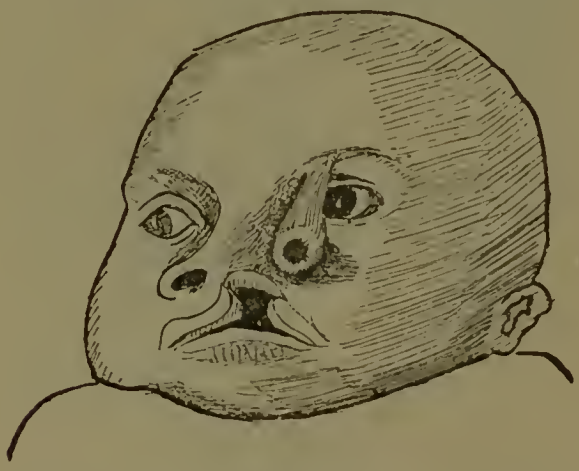

Fig. 12.—Lateral nasal proboscis (after Kirchmayer).

of the nasal cavity is apparently derived from the normally developed olfactory epithelium and olfactory bulb. The same thing is seen in connexion with the ear; when the eighth nerve is absent the external ear is malformed or absent. The case recorded by Kirchmayer[6] differs from all others: he regards the fold crossing the cleft in the lip (see Fig. 12) as the ala nasi of the left side, and the proboscis as a supernumerary structure, which therefore does not represent the left half of the nose. In all other cases the proboscis was the sole representative of the corresponding half of the nose, and it is possible that this is also the condition in Kirchmayer's case. The isolated development of half of the nose as a proboscis does not give rise to a gap or fissure on the face; the maxillary process fuses with the nasal processes of the opposite side (see Fig. 12), thus covering the area normally occupied by the missing half of the nose.

Malformations of the Nasal Cavity.

In Fig. 13 is shown the preparation (No. E 169 in the college collection) of a nose of a newly-born child prepared by Mr. Shattock to show the normal development of the little mucus pocket which represents Jacobson's organ at birth. The pocket contains a small isolated area of the olfactory epithelium vestigial in man, placed on the septum near the anterior nares and the naso-palatine canals. Indeed, in many mammals the orifice of the pocket appears on the palate by the side of the incisive papilla, and is probably stimulated by the odour arising from food as it enters the mouth. Occasionally the septal pocket or recess assumes a considerable size. Mr. Hunter Tod showed me such a case in his out-patients at the London Hospital. Mangakis has figured a case in which a pocket passed along the posterior three-fourths of the nasal septum on the left side,[7] its orifice communicating by a perforation in the septum with the orifice of the corresponding pocket situated on the right side of the septum. Ballantyne gives reference to several cases. I need only mention certain other malformations of the nasal cavity. A specimen in the Hunterian Collection of the college shows a perforation of the anterior part of the septum, in the area where Jacobson's recesses open.

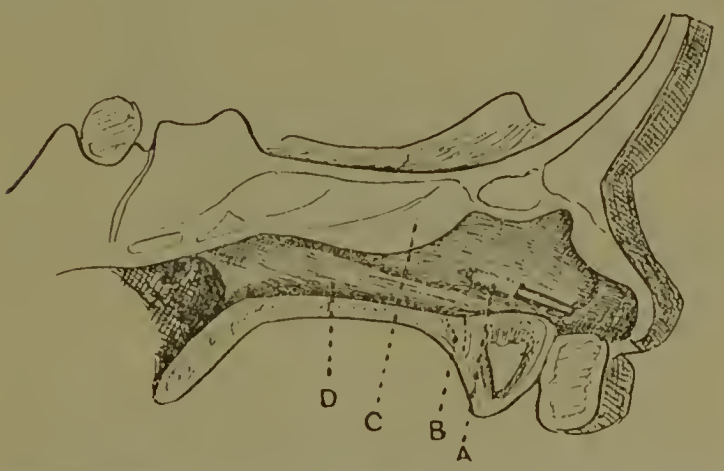

Fig. 13.—Septum of the nose, showing septal recess (Jacobson's organ), in full-time child (Mr. Shattock). A, Septal recess with glass rod inserted at orifice; B, naso-palatine canal; C, middle turbinate process; D, vomer.

Gaup has shown that in the Australian porcupine (echidna) there is an absence of cartilage in the corresponding part of the nasal septum, and in the Australian duckbill the septum is perforated. Absence of and deformities of the nasal septum were mentioned already in describing cleft palate. I regret that there is not a single specimen in the metropolitan museums illustrating occlusion of the posterior nares, a condition which has been frequently described in late years. In a recent paper, Dr. W. G. Porter,[8] in describing a case, mentions that over 100 examples have been recorded. We shall

see that in the condition of cyclops and often in agnathous fetuses there is occlusion of the posterior nares. The explanation offered by Hochstetter is that the nasal cavity in the region of the posterior nares is filled with a solid mass of epithelium in its earlier stages of development; in a later stage, the epithelium breaks down and the nasal cavities then communicate with the naso-pharynx. In some cases the occluding tissue contains

TABLE III.—*List of Specimens showing the Conditions of Cyclopia and Agnathia.*

	A.	B.	C.	Total.
Cyclopia with the nasal processes fused to form a proboscis	6	5	17	28
Cyclopia without a proboscis	—	4	6	10
Cyclopia combined with agnathia	6	—	12	18
Agnathia (ears lateral)	1	1	4	6
Agnathia (oars ventral)	1	3	13	17
Aprosopus (absence of the preauricular part of the face and head)	2	1	8	11
	16	14	60	90
Epignathus	2	5	4	11
Buccal fissures	2	2	2	6
	4	7	6	17

A. Human specimens in the Museum of the Royal College of Surgeons of England.
B. Human specimens in the museums of the metropolitan medical schools.
C. Specimens of vertebrate animals others than man chiefly in the Museum of the Royal College of Surgeons.

bone. The conditions require further examination. Occlusion may occur at the anterior nares. There are two cases in the college collection. The anterior nares are represented by pits, which, as regards depth, correspond to the wings of the nose. Retzius observed that an occlusion of the anterior nares by an epithelial plug occurred during the 3rd, 4th, and 5th months of fetal development. Occlusion of the anterior nares is evidently due to an organization of this plug.

CYCLOPIA.

Although this condition is altogether beyond surgical aid, it is well worthy of study were it only to illustrate the extraordinary manner in which the embryological elements of the face can adapt themselves to an altogether new combination, and produce a structure which is at once malformed and yet finished. Table III shows the great wealth of material which has been collected to illustrate

tho condition; the table aiso shows tho groups into which I have arrranged the material at my disposal, but it would require a series of demonstrations to deal with the various conditions fully. Figs. 14, 15, 16, and 17 will serve to give a general idea of the lesions represented in the various groups of cases included in Table III. Fig. 14 represents a specimen in which the nasal cavities and all the parts pertaining to the nasal cavities are absent, so that the orbits and eyelids and eyeballs fuse in the middle line of the face. There is no apparent trace of the nasal processes in the specimen figured. On the other hand, the maxillary processes are fully formed and meet together. There is also present the condition known as agnathia; the mandible is represented by a mere vestige. The mouth is a minute orifice on a conical process formed out

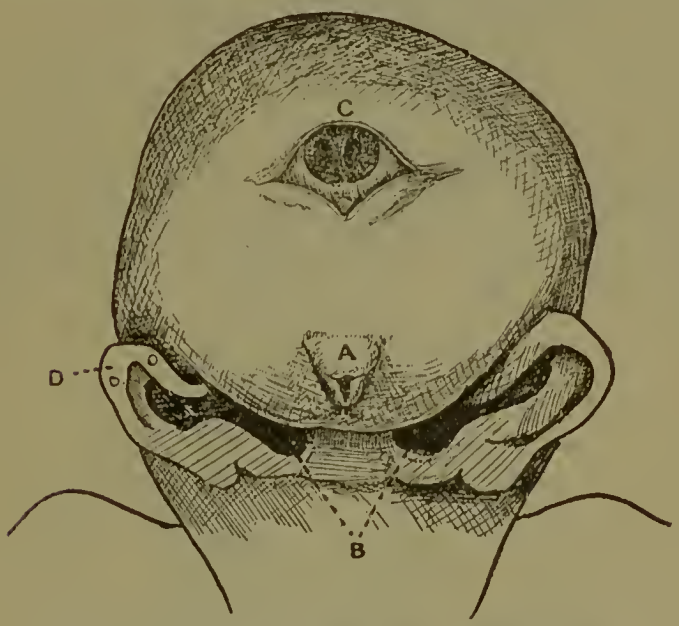

Fig. 14.—Hoad of fotus, showing cyclopia combined with agnathia. A, Proboscis-like mouth; B, external auditory moatus; C, combinod eyeballs; D, right car, showing two fistulae.

of the lips. The external auditory meatus of the ono side almost meets that of the other. In the specimen (No. 263A, Royal College of Surgeons Museum) portrayed, of a child in the seventh month of development, the mandible and nasal processes, which form so large a part of the human face, have been blasted at an extremely early stage of development, and yet the maxillary processes have escaped and attempted to cover up the deficiency. In Fig. 15 is shown a section of the head of a cyclops child (No. 228, R. C. S. Museum) in which the mandible is fully developed, and the nasal processes are represented by a proboscis projecting over the median eye. The *basis cranii* in front of the pituitary body is absent, only that part of the skull base is present which is

developed in connexion with the primitive body axis, the notochord. The optic foramina and sphenoidal fissures form one opening. The optic nerves are fused, and in such cases Mr. Stephen Mayou[9] found no nervo fibres developed within them. The olfactory lobes and nerves aro absent, and the proboscis is made up of the cartilaginous olfactory sense capsules, which enclose a pocket of mucous membrane representing the fused olfactory pits. We are dealing here with an atrophy of all those parts pertaining to the prechordal part of the skull

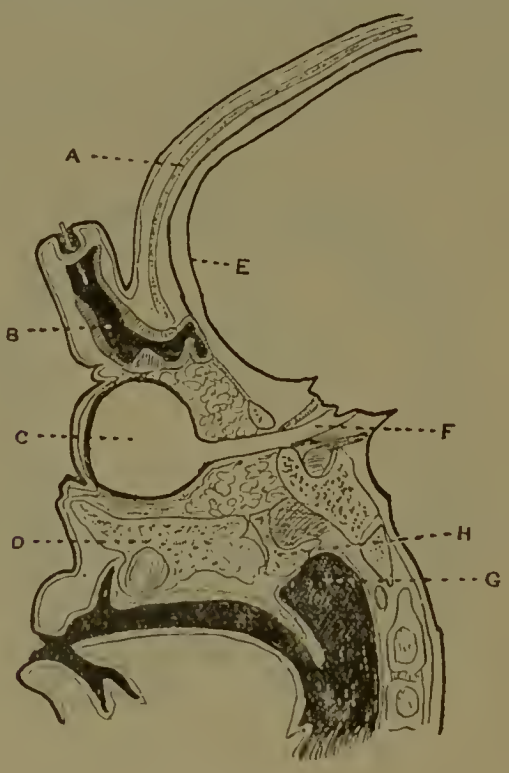

Fig. 15.—Mesial section of head of cyclops. A, Frontal bone in forehead; B, proboscis with capsule of cartilago forming inner wall of cavity; C, fused oyeballs; D, superior maxilla; E, fused anterior ends of corebral vesicles; F, fused optic nerves; G, Eustachian tube opening in naso-pharynx; H, palato bone.

a part which we have reason to believe is of less antiquity than the chordal part. Fig. 16 shows tho formation of the single orbit. All the membrane-formed bones, developed in connexion with the cranial cavity and with the maxillary processes, are present, but the bones formed in connexion with tho nasal processes are absent. The orbital plates of the superior maxillae and palate bones meet to form a complete and finished floor for the orbit. Although the permaxillary element is absent, the palate is neatly formed, and carries no incisor teeth, only the canine and milk molars (see Fig. 17). The position of the posterior nares

is occupied by the vertical plates of the palate bone, which meet in the middle line, there being of course no vomer and no septum. As may be seen from Fig. 15, the naso-pharynx is formed and receives the Eustachian tubes.

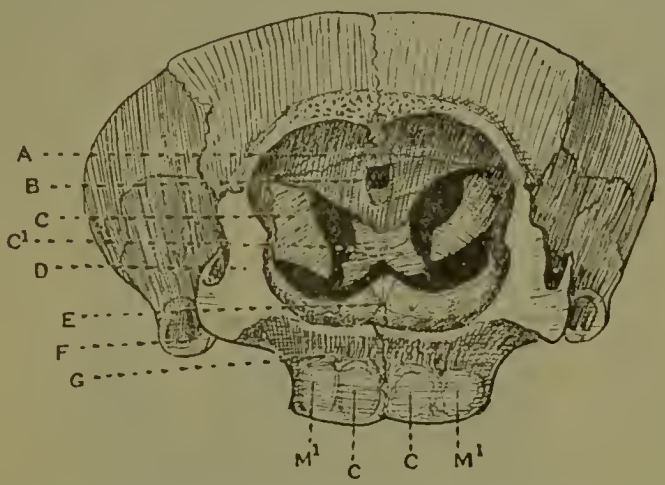

Fig. 16.—Facial part of the skull in a cyclops. A, Orbital plate of frontal; B, foramen for combined optic nerves between small wings of sphenoid; C, great wings of sphenoid: C^1, basi-sphenoid; D, malar; E, orbital process of palate bone; F, annulus tympanicus; G, superior maxilla.

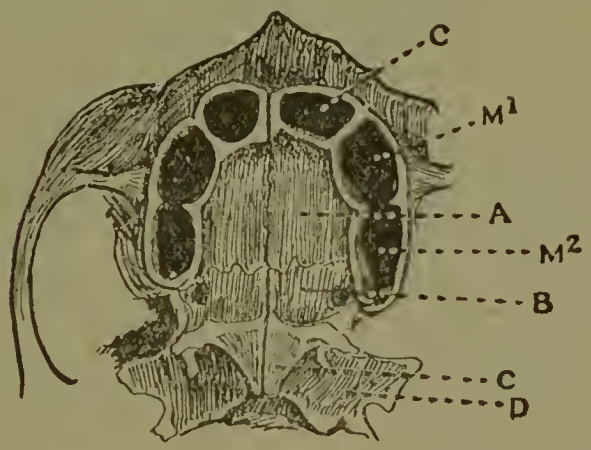

Fig. 17.—Palate in a case of cyclops. C, Socket for canine tooth; M^1, M^2, for first and second milk molars; A, palatal plate of maxilla; B, horizontal plate of palate; C, vertical plate of palate; D, external pterygoid plate.

Agnathia.

The condition of parts in agnathia are shown in Fig. 18. There is, in this case, a total separation of two embryological parts—the stomodaeum or primitive mouth and the pharynx (developed from the foregut), which normally become continuous before the end of the third week of development. In the figure the cavity of the stomodaeum is exposed from below, and shows the palate in a complete form on the roof of that cavity.

There is no trace of the lower jaw, but a mass of fibro-muscular tissue represents its musculature. The lobules of the ears and the tympanic rings are fused on the basal surface of the skull; the malar bones and zygomatic processes occupy a position in the ventral aspect of the skull. The auditory passages open into the pharynx, which contains the tongue. The tonsil lies just above the hyoid bone (see Fig. 18), and over the mylo-hyoid and hyo-glossus muscles.

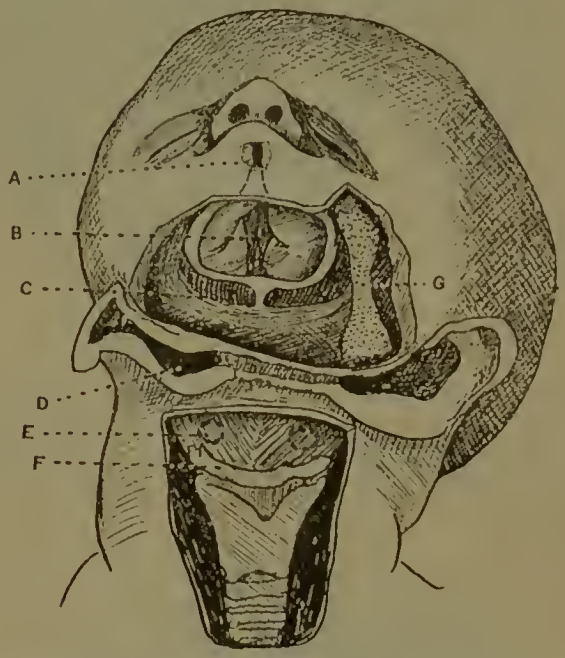

Fig. 18.—Head of full-time child, showing the condition of agnathia. A, Lips and mouth; B, buccal cavity (stomodaeum) exposed from below; C, fibro-muscular mass representing muscles of the lower jaw; D, external auditory meatus; E, mylo-hyoids, with tonsils and tongue lying deep to them; F, hyoid; G, bar of bone representing zygomatic process.

In the conditions just described the nasal or the mandibular elements of the face may be absent, or both may be wanting, but in the last group given in Table III—the condition known as "aprosopus"— the maxillary as well as the two other elements are absent, and the animal appears to terminate in a pharynx and ears.

Epignathus.

The illustration I give to represent this condition is taken from the paper on epignathus by Dr. Alex. Low,[10] in which the fullest and most accurate account of this condition is given. The specimen in the college collection, the two at St. Bartholomew's Hospital, and another at St. George's Hospital, are replicas of the specimen described by Dr. Low. Two others, one in St. Mary's Hospital and another at Guy's Hospital, differ in that the tumour, instead of projecting through the widely opened mouth, protrudes on a distorted face. The epignath may form quite a small tumour, such as represented by

tho specimen recently presented to the college collection (No. 5413) by Mr. Bland-Sutton. The condition is also represented in the college collection by two examples from the pig and two from kittens. The epignathous tumour mass represents a twin embryo, which, owing to the position and conditicn of the development, has to derive its blood supply from the head of the host fetus. The stalk of the tumour is attached to the base of the skull of the host fetus in the region of the basi-sphenoid—the region wherc the notochord, or primitive axis, ends (Fig. 19). When the face is developed the attachment of the epignathial twin becomes included in the septum of tho

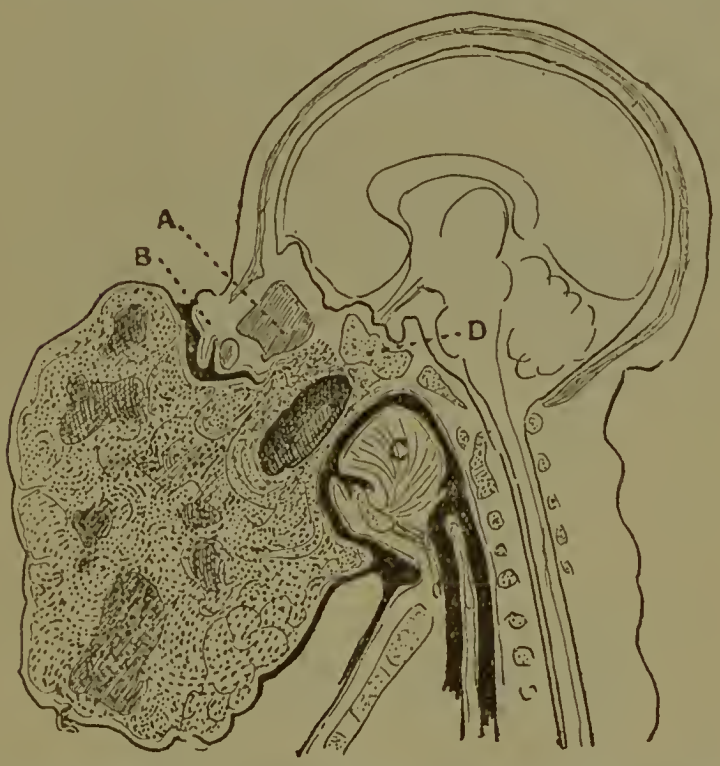

Fig. 19.—Dr. Alex. Low's case of epignathus. A, Nasal septum; B, nose, upper lip, and palate: C, tongue and lower lip; D, the basi-sphenoid.

nose and palate. The size and differentiation of such tumour masses vary widely. Fuller information regarding the nature of epignathous tumour will be found in Principal Windle's paper.[11]

Buccal Fissures and Allied Conditions.

Occasionally clefts or open fissures are found on the cheeks of newly-born children passing from tho angle of the mouth outwards towards the ear, which they may reach, as in a case recently published by Dr. Edgington, or they may stop short at the anterior border of the masseter muscle. Occasionally, as in cases noted by Ballantyne, theso fissures run from some point on the

upper lip and end in the lower lid of the eye. The fissures running outwards from the angle of the mouth, of which I have seen only two in London, one of which is now in the college collection, are usually supposed to represent an imperfect fusion of the mandibular and maxillary elements of the face. I am disposed to think—and the evidence on which I base my inference will be given more fully when I come to deal with supernumerary auricles—that these clefts represent not an arrest of fusion of normal elements, but fissures caused by bands which are supposed to be amniotic in origin and are apt to be formed, during morbid conditions of development, between exposed mucous surfaces such as the lips and ear, lips and eye, nares and exposed meninges. A true condition of macrostomia—of persistence of the fissure between the mandibular and maxillary processes in the human subject—is shown in Fig. 20. The external

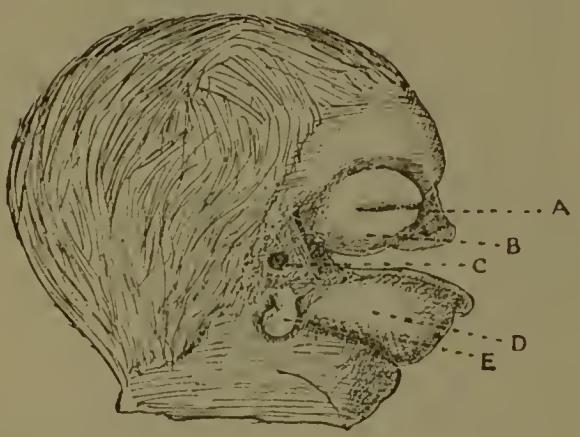

Fig. 20.—Head of human fetus showing a mandibular cleft, absence of the parts developed from the mesial nasal process and eye deformity of the external ear. A, Maxillary process; B, eyeball; C, external auditory meatus; D, mandible; E, auricularappendix.

auditory meatus is present, but the external ear is represented merely by a cutaneous tag below that orifice. The orifice of the mouth extends almost to the meatus. The maxillary processes, owing to an arrest in their growth, have met internal to the orbits, thus leaving the eyeballs on the upper border of the buccal cleft. I have seen this condition in four specimens, two of them being human fetuses, and the other two malformed lambs.

References.

1 Walter L. Woolcombe, *Lancet*, 1905, i, p. 357. 2 R. C. Dunn, British Medical Journal, 1909, i, p. 723. 3 G. Stanguleanu, *Archiv. d'Ophtal.*, 1900, vol. xx, p. 141. 4 F. von Winkel, Sammlung, *Klin. Vorträge*, 1903, Nos. 373, 374. 5 Alex. MacLennan, British Medical Journal, 1903, ii, p. 1581. 6 L. Kirchmayer, *Deut. Zeit. für Chirurgie*, 1906, vol. 81, p. 71. 7 Maugakis, *Anat. Anzeiger*, 1902, vol. xxiii, p. 106. 8 W. G. Porter, *Edin. Med. Journ.*, 1906, vol. xix, p. 129. 9 Stephen Mayou, *Trans. Ophthalmolog. Soc.*, 1906, vol. xxvi, p. 267. 10 Alex. Low, *Studies in Pathology*, edited by Wm. Bullock, Aberdeen, 1906. 11 B. C. A. Windle, F.R.S., *Journ. of Anat. and Physiol*, 1899, vol. xxxiii, p. 277.

III.

Maldevelopment of the External Ear.

In Figs. 21, 22, and 23 are represented the more common forms of accessory auricular appendages. Fig. 21 is from one of the five examples in the college collection. There is here a very distinct arrest in the closure of the first visceral cleft on the sides of which the external ear is formed. The tragus, antitragus, and meatus are pulled towards the chin. The condition was associated with double cleft of the palate and absence of the auditory nerve. In all the three specimens figured the tragus is subdivided, each half projecting as a separate process to a varying degree. A process derived from a subdivision of the tragus is the most common form of auricular appendage. An appeal to comparative anatomy has provided no explanation to the condition. Another very common form of appendix is that shown in Fig. 22, c. It projects from the skin immediately in front of the ascending crus of the helix; when dissected its base is found to be connected with the ascending crus of the helix and to represent the spina helicis, which is usually buried in the subcutaneous tissues, but here projects on the surface. The pendulous process on the cheek (see Fig. 23) is also a form of auricular appendix. There are three specimens in this museum, and I have seen two others. The cheek appendages occur along the line marked in Fig. 23, a line represented by a fissure in cases such as those published by Mr. J. H. Morgan[1] and Dr. Edginton.[2] They are pendulous bodies, consisting of a fibrous core covered by skin. In two of our specimens we found a slight cartilaginous rod in the subcutaneous tissue at the point of attachment of the appendix, passing from the base towards the ear, showing that the cheek processes are really detached elements of the ear. As to the meaning of auricular appendages, I am altogether uncertain; all the specimens in museums show that they are associated with grave malformations, such as club-foot, cleft palate, atresia ani, spina bifida; the cause of the one condition is evidently the cause of the others. Auricular appendages, if they occur alone, are signs of an inherited tendency to grave malformation. The fissures which pass from the angle of the mouth to the ears, the processes which occur on the cheek, and the ventral extension of the tragus (Fig. 21)

are probably caused, as Ballantyne has inferred, by the occurrence of amniotic adhesions.

Fistulae connected with the external ear, which were first described in this country by Sir James Paget,[8] are very imperfectly represented in our museum. There is evidently a relationship between auricular fistulae and auricular appendages, for I noted that one of the commonest sites for their occurrence is in front of the ascending crus of the helix, the point at which a depression or fistula would be formed if a prehelicial appendage became submerged during development (see Figs. 21, 22). The fistula which is found on the ascending helix occurs at the point where two tubercles fuse to form the anterior or mandibular

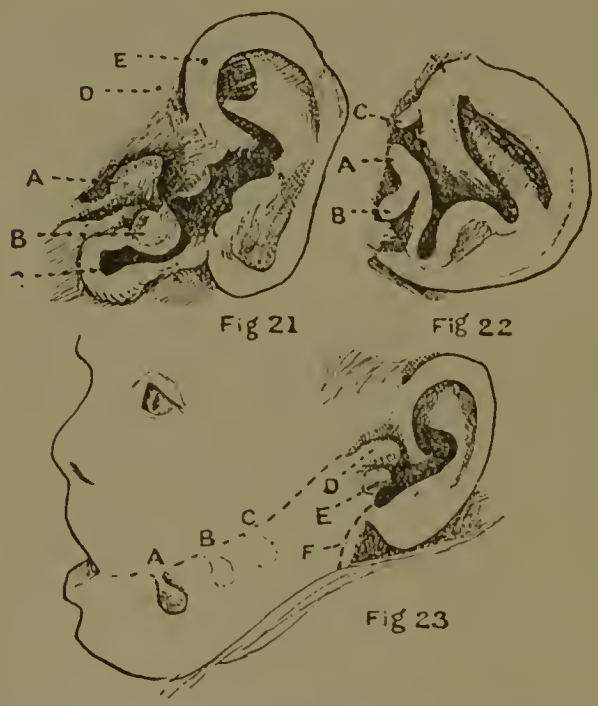

Fig. 21.—Ear of child with cleft palate. A, B, Appendages formed out of the tragus; C, meatus dragged downwards and forwards; D, E, positions of auricular fistulae. Fig. 22.—Ear of an anencephalic fetus, showing A, B, appendages developed from tragus; C, from spine of helix. Fig. 23.—A, B, C, Position of cheek appendages, along the position of a bucco-auricular cleft; D, E, appendages of the tragus; F, position of first visceral cleft (from a photograph in the possession of Dr. William Wright).

part of the helix. When one remembers that the external ear is formed out of six tubercles which fuse together across the first cleft, one wonders that gaps or fistulae are not of more common occurrence.* The external auditory meatus is a secondary formation of the first cleft. It is at first a solid plug of epithelium which afterwards (ninth

* In 50,000 school children Dr. Francis Warner observed 33 with auricular appendages. In 2,000 people, Urbantschitsch (quoted by Pagot) saw 12 with auricular fistulae.

week) becomes canaliculized. A specimen in the college collection shows an arrest in the opening up of the meatus, while the external ear is normally formed. In this case, too, the eighth nerve is absent, and it is observed that malformation of the external ear is frequently attended by a degree of deafness. In the two human specimens in the college museum, with absence or grave arrest of the external ear, there is absence of the auditory nerve.

Dermoids occur in connexion with the first visceral cleft. There are three specimens in the college collection, two of them being retro-auricular and one pre-auricular. I noted two specimens in school museums in which there was a retro-auricular fistula or depression; it is probable that dermoids are formed in connexion with such recesses, which are of uncertain origin.

TABLE IV.

	A.	B.	C.	Total.
Absence, more or less complete, of external ear	2	2	2	6
Accessory appendages of ear	5	2	0	7
Auricular fistulae	—	3	0	3
Cervical appendages	1	—	2	3
Auricular dermoids	3	1	0	4
Auricular fistula with teeth	—	—	2	2
Malformation of tongue	2	1	1	4
Sublingual dermoids	4	1	0	5
Cysts of cervical sinus	12	6	—	18
Cysts or tumours connected with median thyroids	3	2	—	5
Cysts from lateral aberrant thyroids... ...	1	—	—	1
Congenital tumours of the neck	5	2	—	7

A, Specimens (human) in the Museum of the Royal College of Surgeons.

B, Specimens (human) in the museums of metropolitan medical schools.

C, Specimens from domestic animals, chiefly in the Museum of the Royal College of Surgeons.

Cervical Appendages.

The only representative of these abnormal structures in metropolitan museums is a specimen (535) presented to the college collection by Mr. Frederic Eve. He removed it from the neck of a boy, its situation being near the mid-point of the anterior border of the right sterno-mastoid. The lamina of cartilage which supported the process was continuous with a small irregular bar of bone. Mr. Bland-Sutton[4] states that the cartilage is elastic in nature, and that the appendix is usually associated with a cervical fistula. On the other hand, Rieffel,[5] who has collected

37 cases, found that a fistula was never associated with the appendage. In the college collection there are specimens representing the cervical appendage in the pig and goat. It is usually believed that cervical appendages are really auricles developed in connexion with the second cleft. We know that there is a tendency in vertebrates for a segment of the body to assume the characters of an adjoining segment; the last lumbar vertebra, for example, may assume the characters of the first sacral. Here we may have a manifestation on the part of the second cleft to assume to some degree the characters peculiar to the first. There is a difficulty in this assumption which will

Fig. 24.—Ear of ewe showing accessory ostium (Preparation 531, presented by Mr. Bland-Sutton). A, Incisor like tooth; B, ostium distended by rod; C, external auditory meatus.

be dealt with when speaking of cervical fistulae, namely, the second cleft has no separate opening on the neck; it opens in common with the other clefts through the cervical sinus.

Accessory Ostia.

In Fig. 24 is represented the external ear of a sheep, given to the college museum by Mr. Bland-Sutton, who has also described and explained the condition shown by the specimen. Below the ear is a small meatus-like opening on the anterior margin of which is implanted a tooth, apparently an incisor. The abnormal opening may lead into the pharynx or end blindly. Although situated so closely to the ear there can be little doubt

the opening really represents an accessory mouth. The duplication of the hind gut, and formation of two anal openings is a well known condition; duplication of the foregut leading to the formation of a double mouth is also well known. One of the mouths may undergo retrograde changes or imperfect development, and thus come to assume the form seen in these subauricular ostia.

Malformation of the Tongue.

Amongst the specimens at my disposal there are four —three of them in the college collection—which show malformation of the tongue. The head of a lamb (No. 186) shows a most interesting anomaly. On one side the lower

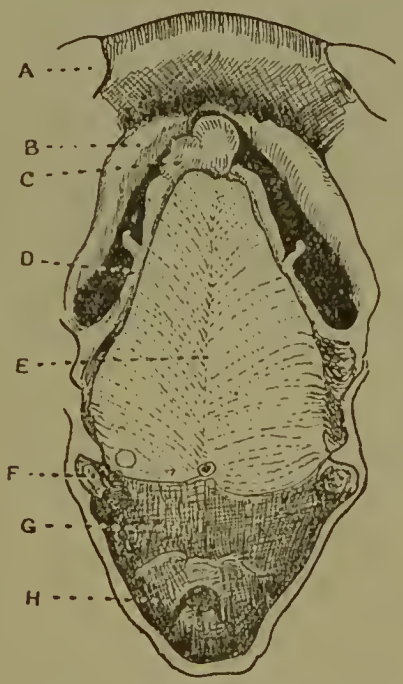

Fig. 25.—Congenital malformation of the tongue. A, Lower lip; B, gums; C, glandular mass distending frenum and extending backwards in paralingual fold (D); E, buccal part of tongue, developed from first arch; F, tonsil; G, pharyngeal part of the tongue, arrested in growth, developed from second and third arches; H, entrance to larynx and rudimentary epiglottis.

jaw and all the parts developed in connexion with it are absent, so that the buccal cavity and tongue are fully exposed on the cheek. The basal part of the tongue—the part behind the foramen caecum and V-shaped sulcus —is fully formed. That part is formed out from the second and third arches, but the anterior or mandibular part of the tongue is only formed on the sound side; on the side of the defect the corresponding half is absent. The

specimen demonstrates two things: (1) That even the anterior part of the tongue is bilateral in origin; (2) that the anterior part of the tongue is entirely derived from the mandibular arch of the embryo, for in the specimen under discussion the right half of this arch and all the structures derived from it are absent. The two other specimens of lingual malformations are to be found in the pathological series of the college museum, where they are classed as tumours of the tongue (Nos. A 2271 and A 2271a); they are exactly of the same type; one was obtained from a deformed, the other from an anencephalic child. One is represented in Fig. 25. At first sight it appears as if the anterior part of the tongue were absent, but such is not the case; it is the basal part of the tongue, the part behind the V sulcus and derived from the second and third visceral arches, which is arrested in development. Combined with this there are two other anomalies: (1) The submaxillary gland has been arrested in its development, and forms a clustered glandular mass within the frenum and along the side of the tongue; (2) Meckel's cartilage has become ossified into a chain of bones, the chain ending behind in the malleus. The sublingual gland is normally developed; the sulcus or depression which is formed in the floor of the primitive mouth so as to cut away the submaxillary gland from the tongue has never opened out. There is in these cases a real condition of tongue tie, the frenum being distended by the glandular mass within it. There is a close relation between these specimens and the cases described by Mr. Barling,[6] Dr. G. W. Griffith,[7] Dr. Kettner,[8] and a case I saw at the London Hospital in a girl aged 2 years. In all of these cases the anterior part of the tongue is imperfectly separated from the floor of the mouth, and in two of them the anterior part of the tongue seemed to be divided, the line of division being occupied by a median mass, which I infer contained muscular and glandular tissue representing submaxillary gland. It is also possible that the median mass may represent the primary element described by His as the tuberculum impar, for Kallius[9] has shown that only a small part of the tongue arises from that element, the major part being formed from lateral elements which grow up from the floor of the mouth and submerge the primitive median element. The fourth specimen on my list (see Table I) is a child in the Museum of St. George's Hospital, which shows a median cleft of the lower lip and jaw, the cleft extending to, and involving, the apex of the tongue.

Sublingual Dermoids.

In the mass of glandular tissue lying by the side of the tongue, in the specimen shown in Fig. 25, there is a small cyst with a lining of mucous stratified epithelium, and in the interior of the cyst, débris of epithelial cells. The cyst has all the characters of a dermoid I have spoken of the downgrowth of a ledge of epi-

thelium which occurs in the floor of the mouth on each side of the tongue, and thus separates off the submaxillary glandular area. It seems highly probable that the cyst alluded to above is a product of the paralingual epithelial downgrowth, and that sublingual dermoids are derived from this downgrowth. There are four specimens of sublingual dermoid cysts in the college museum. Such cysts have been fully described by Mr. Butlin[10] and by Dr. Edginton.[11]

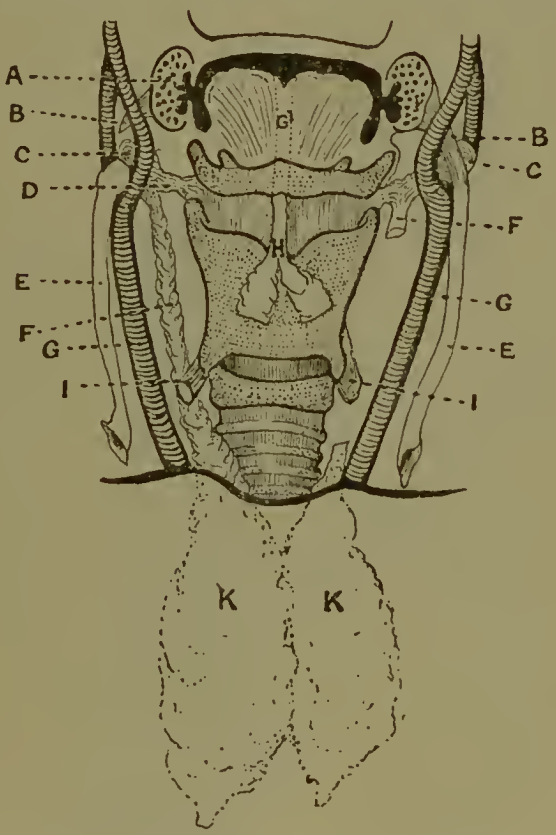

Fig. 26.—Diagram to show the position and connexions of the various embryological remnants in the neck (Dr. S. J. A. Beale). A, Tonsil; B, remnant of tonsillar sac (from second cleft recess); C, carotid body between external and internal carotid arteries; D, stalk of thymus; E, cervical sinus; F, cervical thymus; G, common carotid artery; G, suprahyoid part of median thyroid; H, infrahyoid part of median thyroid, dividing; I, stalk of lateral thyroids; K, K, thymus in thorax.

Embryological Remnants in the Neck.

Before proceeding to describe the various specimens which represent, or are believed to represent, congenital malformations of the neck, it will be well to enumerate, in the light of recent embryological researches by Hammar[12] and of Henry Fox,[13] the various vestigial structures which may assume a pathological form during the course of development and growth. These elements are

shown diagrammatically in Fig. 26. They arise in connexion with the first, second, third, and fourth visceral clefts, the last two being clefts in which gill formations are developed in fishes. From the first cleft arises the median thyroid, the element which forms the isthmus and nearly a half of each lateral lobe of the thyroid in man. The stalk of this element normally disappears, but parts may persist and form tumours or cysts in the line of its formation (Fig. 26). The tonsil is developed at the pharyngeal orifice of the second cleft, but the fundus or

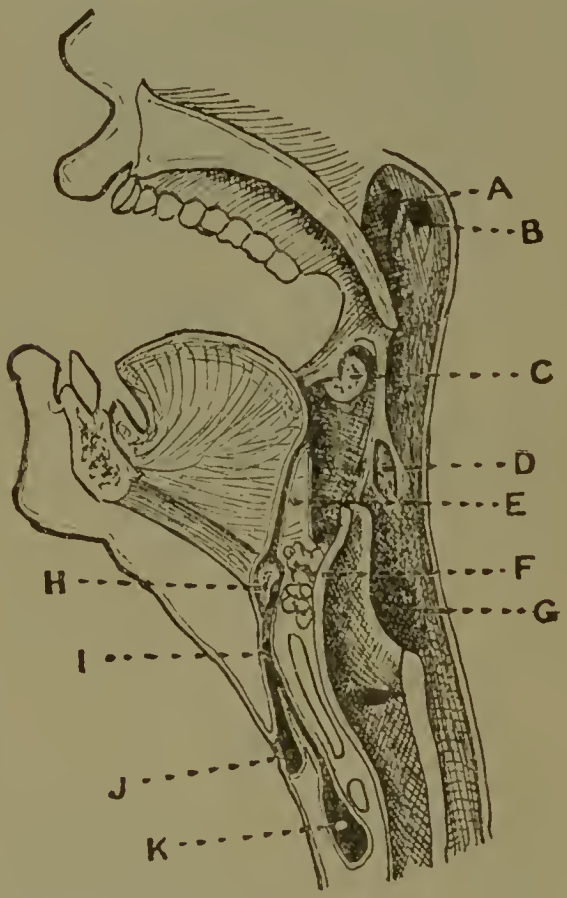

Fig. 27.—Diagram showing the position of the visceral clefts in the human pharynx. A, Eustachian tube (first cleft); B, lateral recess of pharynx; C, tonsil (second cleft); D, position of third cleft, with tonsillar body; E, suprahyoid part of median thyroid stalk; F, retrohyoid mucous glands; G, pyriform fossa (fourth cleft); H, hyoid; I, infrahyoid part of median thyroid stalk; J, right division of median thyroid, forming cyst and fistula; K, left division of median thyroid, forming isthmus.

outer part of the pocket (see Fig. 26) may persist.[14] In the figure this pocket ought to be shown passing between the lesser and greater horns of the hyoid bone (second and third arches), and ending in the fork between the external and internal carotid arteries. From the third

cleft, represented by the thyro-hyoid space, arises the thymus and carotid body; the stalk atrophies in man as far down as the lower pole of the thyroid body, but it may persist, as is normally the case in the pig, and is occasionally the case in newly born children.[15] From the fourth cleft (represented in the interior of the pharynx by the pyriform fossa; see Fig. 27) arises the lateral thyroids and the parathyroids. The stalk of the lateral thyroid buds becomes reduced to a fibrous cord which emerges from the larynx behind the inferior cornu of the thyroid cartilage (Fig. 26, I), and forms in man a firm ligamentous bond (the suspensory ligament) which binds the thyroid body to the cricoid cartilage, but in carnivora, as may be seen in specimens prepared by the prosector of this college (Wm. Pearson) form free fibrous cords. Besides these four endodermal derivates of the visceral clefts—the stalk of the median thyroid, the fundus of the tonsillar clefts, the cervical stalks of the thymus, and the stalks of the lateral thyroids—there is also a fifth element, a derivative of the covering or ectoderm of the embryonic gill area, which may persist and give rise to new formations. That is the cervical sinus (see Fig. 26, E). We see repeated in the human embryo by this sinus, a modification of the gill chambers with which we have long been familiar in the air-breathing fishes—the Dipnoi—and in the more primitive amphibia. In these vertebrates the gill clefts open on the surface of the neck by a common chamber; there is now no doubt that the cervical sinus of the human embryo represents that common gill chamber. During the fourth and fifth weeks the endodermal pockets which give rise to the tonsil, thymus, and lateral thyroids abut on, and are attached to, the fundus of the cervical sinus. The lateral thyroid buds are the first to lose their connexion with the sinus, and the tonsillar pockets the last, because their attachment is the most firm and extensive. There is an especially close connexion between the cervical sinus and the carotid body—a derivative of the third cleft. The fundus of the cervical sinus persists in the mammalian embryo long after its orifice on the neck is closed and in the mammalian embryo becomes a temporary thymus-like body; lymphoid tissue develops in its wall. The question naturally arises: Why is the cervical sinus drawn out into a long fistulous passage (Fig. 26), and why is its orifice on the anterior border of the sterno-mastoid usually near the upper border of the sternum? The answer is to be found in the remarkable change undergone by the neck towards the end of the second month of development. Up to that period, although the cervical part of the spinal column is developed, the chin rests on the breast, the neck being indicated merely by a transverse depression or groove in which the cervical sinus opens on each side (Fig. 28). At the end of the second month and commencement of the third the head becomes extended so that the chin is lifted off the heart and thorax and a neck becomes demarcated. It is not the heart that has receded from the head, but the opposite.

If the surface connexion of the cervical sinus has remained unbroken, then the sinus becomes drawn out into a long tubular structure along with the carotid arteries and trachea. The orifice of the sinus usually occurs on the anterior border of the sterno-mastoid an inch or rather more above the sternum, but under certain circumstances the orifice may be drawn up during extension of the neck to occupy a higher position. Cervical appendages occur at the orifice of the sinus.

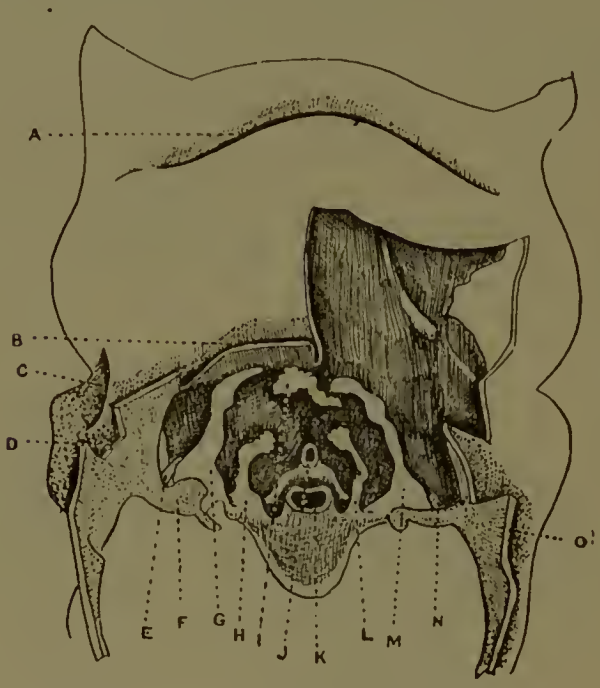

Fig. 28.—Reconstruction of neck of pig embryo, showing the development of the thyroid, thymus, and cervical sinus (after Henry Fox). A, Mouth; B, groove between mandible and thorax; C, external auditory meatus; D, cervical sinus; E, epidermal pocket from cervical sinus to tonsillar recess; F, epidermal pocket to thymus outgrowth; G, M, thymus outgrowths; H, L, lateral thyroids; I, median thyroid; J, trachea; K, oesophagus; N, epidermal process to tonsillar outgrowth; O, cervical sinus.

Cervical Cysts.

There is not, as far as I am aware, a single specimen in metropolitan museums representing a cervical sinus and fistula *in situ*, but, on the other hand, specimens of excised cervical cysts are numerous, two of which are said to have had a fistulous opening. From their structure and relations, it is evident that the majority of these are derived from the cervical sinus. They are sometimes described as dermoids, sometimes as sebaceous cysts, and sometimes as lymphoid cysts. I exclude those cysts which contained grumous matter or blood, because such specimens as have been minutely examined appear to be derivates of the thyroid—cysts formed in aberrant thyroids. Of the eighteen cysts which I regard as derived in part or entirely from

the cervical sinus, twelve are in the college museum. A specimen removed from the neck, opposite the thyroid cartilage, by Sir Felix Semon, may be taken as a typical example. A small bar of cartilage is embedded in its wall—such a cartilage as is occasionally found in the tag at the orifice of the cervical sinus—a product of the operculum which forms the outer wall of the sinus. The cyst is lined by columnar epithelium, similar to that which lines the trachea, but not ciliated. Very little lymphoid tissue is to be found in its wall. The contents are of a milky nature, containing epithelial products. The lining epithelium may be stratified and squamous, or columnar with cilia. Usually these cysts have a close connexion with the carotid sheath, especially near the point of bifurcation of the carotid artery into its internal and external branches. The cervical sinus may fuse and be continuous with the tonsillar recess, so that the cyst wall has both an ectodermal and endodermal lining.

Aberrant Thyroid Tumours and Cysts.

Such tumours and cysts are not well represented in metropolitan medical museums. In the college collection there is a fine specimen presented by Mr. Butlin of a double thyroid tumour, one part of which was removed from the tongue above the hyoid bone; the other part lay below the hyoid. Clearly they were derived from remnants of the median thyroid. Mr. Makins[16] recorded the removal of a thyroid tumour from the posterior part of the tongue in the neighbourhood of the foramen caecum, and recently Dr. Perkins[17] and Dr. T. B. Murphy[18] have been able to bring together about 50 cases of intralingual tumours formed of thyroid tissue, most of them in women. Such tumours frequently occur in the *right* submaxillary region;[19] none are recorded from the same region of the left side. Such tumours apparently represent the right division of the median thyroid. The median thyroids occupy a submaxillary position in the frog. There are two specimens of subhyoid cysts in the college collection, but one of these may be connected with a retrohyoid group of mucous glands which open in the larynx, and were first demonstrated to me by Wm. Pearson, prosector to this college (see Fig. 27). The right division of the median thyroid may be also arrested in development, and form a cyst which afterwards establishes a fistulous opening in the neck.[20] Schrager[21] has recorded 15 cases of cervical cysts derived from the lateral thyroids; they occur beneath the lower third of the sterno-mastoids.

Cysts and Tumours connected with the Tonsillar and Thymic Outgrowths.

There are two remarkable specimens in the museum of Guy's Hospital, numbered 499 and 501. In the first specimen, the pharynx of a man suffocated by a foreign body in the pharynx, there is a recess in the position shown in Fig. 27, containing lymphoid tissue—a form of

tonsil. The recess represents the inner orifice of the third cleft. In the second specimen (No. 501) a cystic tumour, occupying the subparotid region, opens at this depression; its position and connexion make it clear that this tumour is connected with the stalk of the thymic outgrowth. There is a similar tumour in the museum of the Westminster Hospital medical school. Dermoid cysts found in the anterior mediastinum and in the lower part of the neck may be derived from epidermal inclusions, carried down by the outgrowing buds of the thymus from the cervical sinus.

Fig. 29.—Congenital tumour of neck replacing thyroid gland. The right half of the specimen is represented.

Congenital Tumours of the Neck.

Altogether I have seen six specimens of congenital tumour of the neck, four of which are in the college museum. In three of these the tumour is median in position, and has expanded into the neck like a huge goitre (Fig. 29), but in the fourth it has developed to the left side, spreading upwards under the ascending ramus of the jaw, zygoma, and temporal fossa, so as to indent the temporal fossa and compress the brain (Fig. 30), and expand the malar and superior maxilla into thin bony plates which cover the tumour. In none of these specimens is any trace of the thyroid to be found; the tumour is the thyroid. On section these tumours are seen to be multiloculated and solid. On microscopic examination they are found to be composed of compressed cysts lined with columnar epithelium. At the bases of the columnar epithelium a layer of lymphoid tissue usually is

found; indeed, a large part of the tumour is lymphoid in nature. In the septa between the spaces are found islands of cartilage. How the thyroid body of the fetus comes to assume such a structure and size is an enigma. The tumours resemble congenital sacral tumours, and in many ways have much in common with the tissues which form gills.

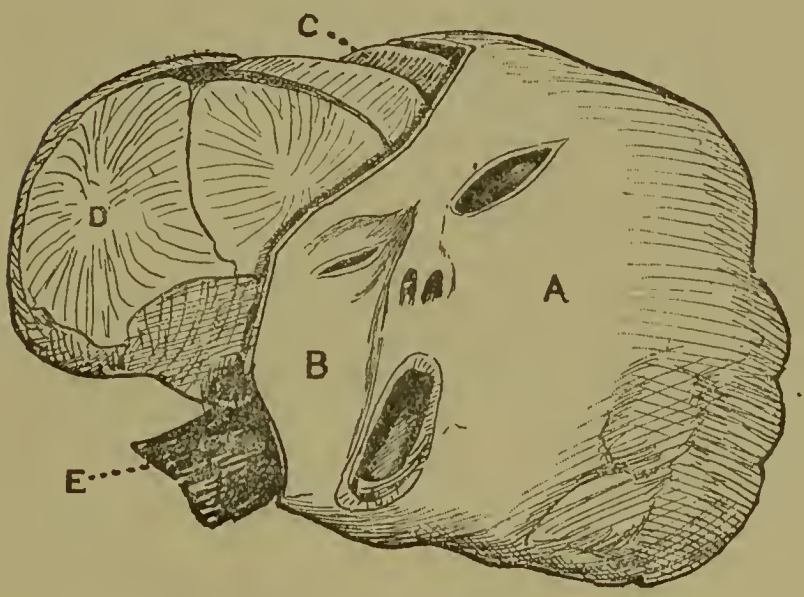

Fig. 30.—Congenital tumour of the neck extending into and filling the left temporal fossa. A, Left cheek in which the malar and superior maxilla are extended as thin bony plates; B, right cheek; C, bony ridge representing temporal crest; E, spinal column and muscles.

REFERENCES.

[1] J. H. Morgan, *Proc. Roy. Med. Chir. Soc.*, London, 1881, vol. ix, p. 94. [2] Edginton, *Glasgow Medical Journal*, May, 1909. [3] Sir James Paget, *Med. Chir. Trans.*, 1878, vol. 61, p. 41. [4] J. Bland-Sutton, *Tumours, Innocent and Malignant*, 4th ed., London, 1906. [5] M. Rieffel, *Bull. et. mém. de la Soc. de Chir.*, 1904, t. xxx, p. 110. [6] Gilbert Barling, BRITISH MEDICAL JOURNAL, 1885, ii, p. 1061. [7] G. W. Griffith, BRITISH MEDICAL JOURNAL, 1899, ii, p. 273. [8] Kettner, *Charité Annalen*, 1907, Bd. 31, p. 400. [9] Kallius, *Verhand. anat. Gesellsch.*, April, 1908. [10] Butlin and Spencor, *Diseases of the Tongue*, 2nd ed., London, 1900. [11] Edginton, BRITISH MEDICAL JOURNAL, 1904, ii, p. 808. [12] Hammar, *Anat. Anz.*, 1902, Bd. xxii, p. 221. [13] Henry Fox, *Amer. Journ. of Anat.*, 1908, vol. viii, p. 187. [14] See paper by J. Seccombe Hett, *Lancet*, 1909, i, p. 457. [15] Bishop Harman, *Journ. Anat. and Physiol.*, 1902, vol. xxxvi, p. 47. [16] G. Makins, *Lancet*, December 8th, 1906. [17] C. E. Perkins, *Laryngoscope*, 1908, xviii, p. 293. [18] T. B. Murphy, *Journ. Amer. Med. Assoc.*, 1905, vol. xlv, p. 1854. [19] See Schrager. [20] See C. F. Marshall, *Journ. Anat. and Physiol.*, 1892, vol. xxvi, p. 94. [21] V. L. Schrager, *Surgery, Gynaecology, and Obstetrics*, 1906, vol. iii, p. 465.